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## Case Report

# Congenital hypopituitarism: A case report

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### ABSTRACT

Congenital hypopituitarism can be life threatening in neonates while its early diagnosis is a greater challenge for the dentist. Hypopituitarism can result from diseases of the hypothalamus or of the pituitary gland. In adults it is often missed due to its nonspecific symptoms of growth hormone deficiency hence early diagnosis and prompt treatment is necessary. Here we have presented a 9-month follow-up of an 8-year-old child with congenital hypopituitarism and illustrated with short stature and delayed overall development. We are focused on dental management for the challenges associated with the disease and treated the dental caries with restorations and a space maintainer was also given. While at the end of 9 months, positive results were seen clinically as well as radiographically.

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## 1. Introduction

Hypopituitarism refers to decreased secretion of pituitary hormones, which can result from diseases of the hypothalamus or of the pituitary gland. Hypofunction of the anterior pituitary occurs when about 75% of its part is lost or absent. This may be congenital or acquired abnormalities that are intrinsic to the pituitary.<sup>1</sup> According to earlier studies, in India, there is an incidence rate of 12-42 cases per million per year, and a prevalence of 300-455 cases per million had been reported in India,<sup>2</sup> the estimated prevalence of the pituitary disorder in the year 2000 was 4 million (Dr. Kochupillai).<sup>3</sup> The onset of symptoms can be sudden and rapid in which generally growth hormone deficiency occurs initially followed by Luteinizing hormone deficiency, while the deficiency of Follicle-stimulating hormone, thyroid-stimulating hormone, adrenocorticotrophic hormones occurs at a later stage.<sup>4</sup> Growth hormone is essential for growth and development. Its deficiency results in a condition known as pituitary dwarfism which has delayed growth of the skull, facial skeleton and, gives a small facial appearance. The deficiency of growth

hormone (GH) can be measured by an insulin tolerance test in which GH levels lesser than 3 $\mu$ g/L are considered to indicate a partial deficiency. Growth hormone level between 3.0-4.9  $\mu$ g/L indicates a partial deficiency, and GH level higher than 5.0  $\mu$ g/L are considered as normal.<sup>5</sup> Symptoms associated with growth hormone deficiency include lack of vigor, decreased tolerance of exercise, and decreased social functioning, diminished muscle mass, increase fat mass while diminished facial and body hair, and fine facial wrinkles were reported on long-standing hypogonadism due to hypopituitarism.<sup>6</sup> Oral manifestations related with hypopituitarism are delayed eruption rate as well as delayed shedding of deciduous teeth, a smaller clinical crown of teeth, smaller roots with retarded growth of supporting apparatus smaller dental arches, crowding and malocclusion, retarded growth of mandible, fine wrinkles around the mouth and eyes and decrease salivary flow.<sup>7</sup>

In this case report the child was noted with decreased BMI than normal, thin body hair, puffy face, wrinkles around eyes, decrease muscle mass, incompetent lips with, delayed eruption of permanent teeth, and poor oral hygiene maintenance all of which brings our attention toward retarded growth and suggestive of hypopituitary disorder.

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This is a disease which is causing not only systemic but also associated with dental anomalies while in some cases oral involvement precedes other symptoms hence it is important for both endocrinologists, as well as the dentist to communicate to perform diagnostic tests to provide a better treatment plan and monitor the medication along with regular follow-ups, will lead to the success of a multidisciplinary approach for its management.

## 2. Case History

A male child aged 8 years reported to the department of pediatric and preventive dentistry of Army College of Dental Sciences with a chief complaint of pain while eating since last one month and also complaint of delay in tooth eruption of the upper front tooth. (Figure 1)



**Fig. 1:**

### 2.1. Medical history

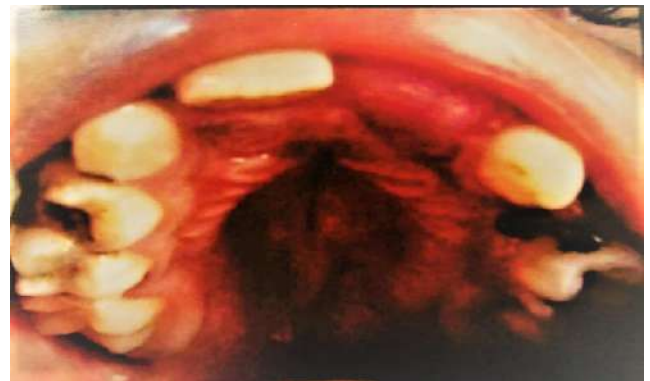
Medical history revealed that patient had, congenital hypopituitarism and is taking human growth hormone injections (Genotropin / Pfizer) from one year of age, the drug dosage was changed according to his body weight. No relevant family history was noted the child's height was 90cm and the weight of the child was 12 kg on the basis of body mass index (BMI) calculated to be 14.8 which was <18.5 and suggestive of underweight.

### 2.2. Extraoral and intraoral examination

Extraorally the child was having a puffy face with thin hairs, deficient nasal bridge, incompetent lips, dry lips (Figure 2), while on intraoral examination reveals inflamed gingiva with calculus, with lower anterior crowding, while Dental caries with respect to 54,55,65 and root stumps with respect to 74,84,85,64 was reported for which intraoral periapical radiograph was taken to diagnose the extent of dental caries and pulpal involvement. (Figures 3 and 4)



**Fig. 2:**



**Fig. 3:**

### 2.3. Diagnosis and treatment plan

On clinical examination and radiographic interpretation, the diagnosis was made i.e. suggested dental caries (54,55,65) and root stumps (75,84,85,64) and delayed eruption with respect to 21. For all the dental caries the conservative approach was opted and the restorations were done with respect to (54,55,65) on the first appointment while on the



**Fig. 4:**

next appointment the root stumps were extracted and the band and loop space maintainer was given. post-operative oral hygiene instructions were given and postoperative medication was prescribed while the patient was kept on a regular follow-up.

#### 2.4. Follow-ups

On a 9 month follow up the 21 was seen clinically erupting while 35 was seen radiographically. (Figure 5)



**Fig. 5:**

### 3. Discussion

Congenital Hypopituitarism can be life threatening in neonates while early diagnosis of which is a greater

challenge for the dentist to alleviate metabolic and developmental consequences of hormone deficiency and to provide a better growth and development of a child.

The pituitary gland is a small endocrine gland that anatomically lies in the hypophyseal fossa/Sella terti /pituitary fossa and controls the secretion of many endocrine glands of the body. The pituitary gland consists of two lobes anterior and posterior lobes. Physiologically the anterior lobe is associated with have ketogenic, anti-insulin, diabetogenic, parathyrogenic, and pancreatotropic activity while the posterior lobe, have been reported with, vasoconstrictive, oxytocic, and antidiuretic activities.

Many studies have shown that either hypopituitarism or hyperpituitarism both had affect the development of oral tissue.<sup>7</sup>

If the hypo functioning of the pituitary gland is seen before puberty it will lead dwarfism which manifest with growth hormone deficiency.<sup>7</sup>

If the deficiency occurs after puberty it affects other endocrine glands also. Some of the common causes of hypopituitarism are associated with pituitary adenoma, Simmonds disease or hypophysial cachexia, and Sheehan's syndrome.<sup>7</sup>

In our case the medical history revealed that patient had, congenital hypopituitarism and was not associated with any other disorder.

The most common feature of Hypopituitarism are the short stature, well-proportioned body, fine hair on the head, wrinkled atrophic skin, and also seen with hypogonadism. While the maxilla and mandible of the patient are smaller than the normal and the face also appears smaller. Permanent tooth delayed in eruption while shedding pattern of deciduous teeth is also delayed by several years and also the development of the roots of permanent tooth appears to be delayed. Because of smaller dental in such patients cannot accommodate all teeth hence results in malocclusion while a complete absence of bud of the wisdom tooth has also been reported.

In the present case report the growth was delayed as the BMI was reported less than normal. Along with the delayed general body growth, short stature, rate of eruption and the shedding time of the teeth is also delayed. While Suspicion of congenital hypopituitarism can be aroused by many things. Prenatally by the family history and ultrasound while postnatal-low T-4 level with normal TSH level, hypoglycemia with or without seizures and jaundice can be seen initially while oral examination for cleft lip and palate and single central incisor should be done.<sup>8</sup>

So it is very important for both endocrinologist and the dentist to communicate with each other to order diagnostic tests, evaluate the results and check, administer and monitor treatment plan and medication designed.<sup>9</sup>

Hypopituitarism is diagnosed by history and physical examination. Along with this two types of blood test are

used to check the deficiency of hormones: basal levels (ACTH, GH) and dynamic levels (LH/FSH, Prolactin, TSH). MRI and CT are also helpful in diagnosing structural abnormalities.<sup>10</sup>

GH involves daily subcutaneous injections or recombinant human growth hormone dosage between 0.16-0.24 mg/kg/week while Glucocorticoid replacement should be begun before thyroid hormone replacement while hydrocortisone dose should be given in a minimum of 3 doses in a day for total of 8 to 12 mg/m<sup>2</sup>/d for maintenance and also levothyroxine is added to attain normal T4 level rather than TSH level. (Higuchi A, Hasegawa Y et al. 2006). While in the present case the child was in prepubertal stage and was having only human growth hormones.

And H. G. Artman and E. Boyden et al. had shown that delay in tooth eruption has been calculated to be from one to three years for teeth that normally erupt in the first decade of life and from one to ten years for teeth erupting in the second decade. Considering which we have kept the child on follow up and the tooth erupted in the oral cavity after 9 months.

#### 4. Conclusion

The very diversity of patients which depends on the age, underlying etiology, extent and duration of the pituitary hormone deficit are commonly seen in clinical practice has also tended to limit systematic investigations of dental development. If at all left untreated later it is seen associated with symptoms like fatigue general weakness, reduced vitality and physical strength, diminished mental ability and ultimately lead to decrease in quality of life

The recognition is often delayed until growth failure develops in the late childhood in spite of telltale sign such as hypoglycemia and jaundice as neonates. Hence developing a suspicion of hypopituitarism based on prenatal and postnatal clues we as a dentists play a key role in diagnosis and early management of pituitary disorder in co-ordination with medical professionals.

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#### 6. Conflict of Interest

The authors declare that there is no conflict of interest.

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